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ORIGINAL RESEARCH

Radiological and clinical features of vein of Galen malformations

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ABSTRACT

Background Vein of Galen malformations (VOGMs) are rare and complex congenital arteriovenous fistulas. The clinical and radiological features of VOGMs and their relation to clinical outcomes are not fully characterized.

Objective To examine the clinical and radiological features of VOGMs and the predictors of outcome in patients.

Methods We retrospectively reviewed the available imaging and medical records of all patients with VOGMs treated at the University of California, San Francisco between 1986 and 2013. Radiological and clinical features were identified. We applied the modified Rankin Scale to determine functional outcome by chart review. Predictors of outcome were assessed by χ^2 analyses.

Results Forty-one cases were confirmed as VOGM. Most patients (78%) had been diagnosed with VOGM in the first year of life. Age at treatment was bimodally distributed, with predominantly urgent embolization at <10 days of age and elective embolization after 1 year of age. Patients commonly presented with hydrocephalus (65.9%) and congestive heart failure (61.0%). Mixed-type (31.7%) VOGM was more common in our cohort than purely mural (29.3%) or choroidal (26.8%) types. The most common feeding arteries were the choroidal and posterior cerebral arteries. Transarterial embolization with coils was the most common technique used to treat VOGMs at our institution. Functional outcome was normal or only mildly disabled in 50% of the cases at last follow-up (median=3 years, range=0–23 years). Younger age at first diagnosis, congestive heart failure, and seizures were predictive of adverse clinical outcome. The survival rate in our sample was 78.0% and complete thrombosis of the VOGM was achieved in 62.5% of patients.

Conclusions VOGMs continue to be challenging to treat and manage. Nonetheless, endovascular approaches to treatment are continuing to be refined and improved, with increasing success. The neurodevelopmental outcomes of affected children whose VOGMs are treated may be good in many cases.

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INTRODUCTION

Vein of Galen malformations (VOGMs) are arteriovenous fistulas hypothesized to arise in the fetal period.[1] VOGMs present antenatally, in early infancy, or in childhood with signs and symptoms ranging from high-output congestive heart failure (CHF) and hydrocephalus to seizures and intracranial hemorrhage.[2 3] Before the advent of surgical and endovascular embolization treatments, VOGMs were universally associated with poor outcomes.[4–7] Historically, cases presenting neonatally had mortality rates of 88–100% and those presenting in infancy or childhood had mortality rates of 40%.[8]

Endovascular embolization techniques have reduced mortality and morbidity in patients with VOGMs. The goals of endovascular treatment vary depending on clinical presentation. Neonates with CHF refractory to medical management are treated urgently to establish hemodynamic stability and to allow for heart and brain development.[9] In neonates without CHF, the goal of treatment is to prevent cerebral venous hypertension, white matter damage, and the "melting brain syndrome". In these cases, treatment is often deferred until the child is aged 5–6 months to allow for greater cerebral maturation and enlargement of arteries usable for therapeutic access.[9 10] Studies have shown a 15% mortality rate in patients treated with endovascular therapy, compared with 76.7% in untreated patients and 84.6% in patients treated with microsurgery.[11–15]

Despite these survival benchmarks, the clinical outcomes of children treated with endovascular techniques are not well characterized. After treatment with earlier endovascular techniques, an estimated 23–70% of children were neurologically normal at short-term follow-up.[11–13 16 17] Previously, a 2003 study at the University of California at San Francisco (UCSF) showed favorable outcomes in 52% of all cases. The median duration of follow-up at that time was 4.5 years (range=0.75–14 years for chart review). Poor outcomes were associated with perinatal presentation, the presence of CHF, and choroidal angioarchitecture.[17] Additionally, in the landmark Hospital Bicetre series of 233 patients with VOGM, studied between 1981 and 2002, 143 (74%) of 193 survivors were reported to be neurologically normal on last reported follow-up.[10] Other studies have also shown encouraging outcomes.[10 14 18]

Thus, we examined children who underwent endovascular treatment of VOGMs at UCSF to characterize the clinical and radiological features of the VOGMs and to determine the predictors of functional outcome. We also aimed to document the advances in endovascular approaches used to treat VOGMs since 1986.

METHODS

Sample

Patients who underwent endovascular treatment of VOGMs at UCSF between 1986 and 2013 were included. The UCSF interventional neuroradiology database was queried for cases involving "vein of Galen malformation" under an institutional review board-approved protocol. Sixty-two cases were identified. Of these, 41 cases were confirmed to have a VOGM based on the criteria defined by Lasjaunias *et al*.[12 19] Twenty-one cases represented non-VOGM diagnoses; often the search term identified VOGM in the patient chart not in the context of the diagnosis. Of the 41 VOGM cases, angiographic images were available for 22. Seven of these cases had digital images on the UCSF Picture Archiving and Communication System (PACS), while 15 cases had images on film. The angiographic images of 19/41 VOGM cases were irretrievable, and these cases were assessed through radiology procedure notes. The demographics and clinical information of the sample are presented in table 1.

Table 1 Patient demographic and clinical information

Demographic factors	No (%)
Gender	
Male	31 (75.6)
Female	10 (24.4)
Age at first diagnosis	
≤10 days	22 (53.7)
11–100 days	5 (12.2)
101–200 days	2 (4.9)
201–365 days	3 (7.3)
>1–2 years	2 (4.8)
>2–3 years	1 (2.4)
Age at first procedure	
≤10 days	12 (29.3)
11–100 days	6 (14.6)
101–200 days	6 (14.6)
201–365 days	4 (9.8)
>1–2 years	2 (4.8)
>2–3 years	2 (4.8)
>3–4 years	0 (0.0)
>4–5 years	5 (12.2)
>5 years	1 (2.4)
Diagnostic modality	
Postnatal CT	12 (29.3)
Postnatal MRI	9 (22.0)
Fetal ultrasound	6 (14.6)
Angiogram	4 (9.8)
Postnatal ultrasound	3 (7.3)
CHF	
Present	25 (61.0)
Absent	12 (29.3)
Seizures	
Present	12 (29.3)
Absent	19 (46.3)
Hydrocephalus	
Present	27 (65.9)
Absent	8 (19.5)
Intracranial hemorrhage	
Present	14 (34.1)
Absent	15 (36.6)

CHF, congestive heart failure.

Imaging evaluation

Two neurointerventional radiologists (SWH and DLC) reviewed the available angiographic, CT, and MRI studies of the cohort to characterize the VOGM imaging features using a standardized record. The diagnostic details, such as the year of diagnosis and the radiological modality employed to make the diagnosis, were recorded. Follow-up imaging on the UCSF PACS and radiology records were also assessed.

Chart review

A single study investigator (MLC) used a standardized data abstraction form to review all available medical records from pediatric neurology and neurointerventional radiology departments. Demographic features, symptoms at presentation, VOGM classification, and other clinical features were recorded. Details of the endovascular treatment and follow-up information were also obtained.

Functional assessment

A single study investigator (MLC) applied the modified Rankin Scale (mRS) to assess the functional outcome of the cases at the last available follow-up.[20] The Social Security Death Index was searched for cases with inadequate chart information.

Statistical analysis

All statistical analyses were conducted using SPSS 21.0 (SPSS Inc, Chicago, Illinois, USA). Summary and frequency statistics were applied to analyze the demographic, clinical, and radiological characteristics of the sample. Two-tailed χ^2 tests were applied to assess the associations between case variables and outcome and clinical and radiological characteristics. χ^2 Tests were also used to assess the differences in endovascular techniques used before and after the arbitrary year 2000. A significance threshold of $p<0.05$ was applied.

RESULTS

Sample demographics and clinical characteristics

Thirty-one male and 10 female patients met the inclusion criteria. The age at first presentation and diagnosis ranged from the first day of life to 2.5 years (median=1 day, range=1 day to 2.5 years). More than 50% of the patients in our cohort were diagnosed before the age of 10 days, and about 7.3% of the patients were diagnosed after 1 year. The age at first angiogram ranged from 1 day to 6 years (median=120 days). Twenty-nine per cent of patients were treated before the age of 10 days, and 24.4% were first treated after 1 year (table 1).

In total, 65.9% of patients presented with hydrocephalus, 61.0% with congestive heart failure, 34.1% with intracranial hemorrhage and 29.3% with seizures (table 1). Postnatal CT was the most commonly used diagnostic modality, followed by postnatal MRI, fetal ultrasound, postnatal ultrasound and angiography (table 1).

Imaging features

The majority of patients had mixed-type VOGMs (31.7%; table 2), followed by the mural-type (29.3%) and the choroidal-type (26.8%). The most common feeding arteries of the VOGM included the posterior and/or anterior choroidal, posterior cerebral, thalamoperforating, anterior cerebral and pericallosal arteries (table 2). Patent falcine veins, absent straight sinuses, sinus stenosis, anomalous dural sinuses and venous enlargement were among the most commonly identified angiographic features (table 2). Fewer cases showed complete limbic arches, connections to the deep

Table 2 Imaging features of the vein of Galen malformations

Diagnostic features	No (%)
Choroidal	11 (26.8)
Mural	12 (29.3)
Mixed	13 (31.7)
Common feeding arteries	
Choroidal artery	28 (68.3)
Posterior cerebral artery	19 (46.3)
Thalamoperforatoring artery	18 (43.9)
Anterior cerebral artery	8 (19.5)
Pericallosal artery	6 (14.6)
Radiological features	
Venous enlargement	22 (53.7)
Anomalous dural sinus	20 (48.8)
Patent falcine vein	19 (46.3)
Sinus stenosis	16 (39.0)
Absent straight sinus	13 (31.7)
Venous ectasia	12 (29.3)
Venous reflux	11 (26.8)
Limbic arch	10 (24.4)
Alternate pathways of venous drainage	9 (22.0)
Connection to deep venous system	6 (14.6)
Occipital sinus	5 (12.2)
Periventricular drainage	5 (12.2)
Venous sinus thrombus or occlusion	4 (9.8)

venous system, alternate pathways of venous drainage, occipital sinuses, periventricular drainage, venous ectasia, venous reflux and venous sinus thrombus or occlusion. Representative imaging findings are shown in figure 1 and online supplementary figure 1. The frequency of imaging features read directly from the angiogram is listed in online supplementary table 1.

Treatment

The total number of endovascular treatments for each case ranged from none to seven, with a median of two treatments (table 3). Only one patient was not treated, and the VOGM spontaneously resolved.[21] One patient received seven total endovascular treatments, with two of them at UCSF.

In the cohort, 27 patients were treated at UCSF before the year 2000, and 14 patients were treated at UCSF in the year 2000 or later. There were no differences in total number treated before and after 2000 (table 3; p=0.219). Transtorcular approaches were not used after 2000. There was no difference between the frequencies of transarterial (p=0.919) or transvenous approaches before and after 2000 (p=0.604).

The embolic agents used in the treatment of the cohort included: coils, silk sutures, N-butyl cyanoacrylate, polyvinyl alcohol (PVA), liquid coils, and platinum wire (table 3). The use of silk sutures differed significantly before and after 2000 (p=0.014). No silk sutures, PVA, or platinum wire were used in treatment after 2000.

Associations between imaging and clinical characteristics

Next, we looked for associations between the imaging and clinical characteristics by stratifying the patients into three groups based on the degree of VOGM venous varix dilatation (mild, moderate, or severe). There were nine cases in the mild group, 11 in the moderate group and four cases in the severe group. We found significant differences in frequency between groups in the presence of venous reflux (p=0.006; moderate > mild=severe), the number of feeding connections (p<0.001; moderate > mild > severe), the presence of seizures (p=0.034; mild > moderate > severe), and the presence of a pericallosal artery feeder (p=0.036; moderate > mild=severe). The severity of the VOGM dilatation was not associated with CHF (p=0.337), hydrocephalus (p=0.523), or intracranial hemorrhage (p=0.060).

χ^2 Tests were also applied to compare the characteristics of the choroidal-, mural- or mixed-type VOGMs. There were 11 cases in the choroidal group, 12 in the mural group, and 13 in the mixed group. Significant differences in frequency were found between groups in the presence of thalamoperforating artery feeders (p=0.014; mixed > choroidal > mural), presence of anterior cerebral artery feeders (p=0.010; choroidal > mixed > mural), and number of feeding connections (p=0.021; mixed > choroidal > mural).

In addition, we explored the relationship between seizures and intracranial calcifications secondary to high-pressure damage. Of the five patients with evident calcifications on imaging, four presented with seizures; the seizure status of the remaining individual was unknown. The embolization procedures for each of these patients were performed at 3 months, 19 months, 2 years, 4 years, and 4 years. Of the five patients, embolization was complete in three, partially complete in one, and incomplete in the remaining one patient. Whether the calcifications and seizures were affected by the embolization procedures is unknown. On neurologic follow-up, however, four of these five patients had developmental delays in motor and cognitive domains after their procedures.

Outcome based on chart review

Median follow-up for chart review was 3 years, range=0–23 years. Of the patients (table 3), 9/36 (22.0% of the total number of patients) are known to have died (mRS=6). In total, 19.5% were neurologically normal on last follow-up (mRS=0). Another 24.4% of cases (mRS=1 and 2) were mildly affected and remained functional. The remaining 22.0% were moderately or severely affected (mRS=3–5). The five (12.2%) remaining patients are known to be living based on the Social Security Death Index. There were no differences in mRS scores between the group treated before and after the year 2000 (p=0.913).

Of the 20 patients who were diagnosed on the first day of life, 11 survived (55%). We compared those who survived with those who died using a χ^2 test on all clinical and radiological features. Venous sinus dilatation (p=0.016) and the presence of a straight sinus (p=0.038) were associated with the patients who did not survive.

A worse final mRS was significantly associated with younger age at first diagnosis (p=0.028), the presence of seizures (p=0.008) and CHF (p=0.044), incomplete or limited fistula thrombosis after treatment (p=0.010), and the use of PVA as the embolic material for endovascular treatment (p=0.015). All nine patients with a mRS of 6 were diagnosed on the first day of life, though three cases also diagnosed on the first day of life had mRS scores of 0, 2, and 4. Final mRS was not predicted by the year of treatment (p=0.151) or angioarchitecture subtype (p=0.534). Death was associated with younger age at first treatment procedure (p=0.017), the presence of CHF (p=0.017) and hydrocephalus (p=0.037), the presence of a straight sinus (p=0.027), the presence of a posterior inferior cerebellar feeding artery (p=0.002), incomplete or limited fistula obliteration (p=0.004), and lack of fistula thrombosis on follow-up imaging (p=0.01).

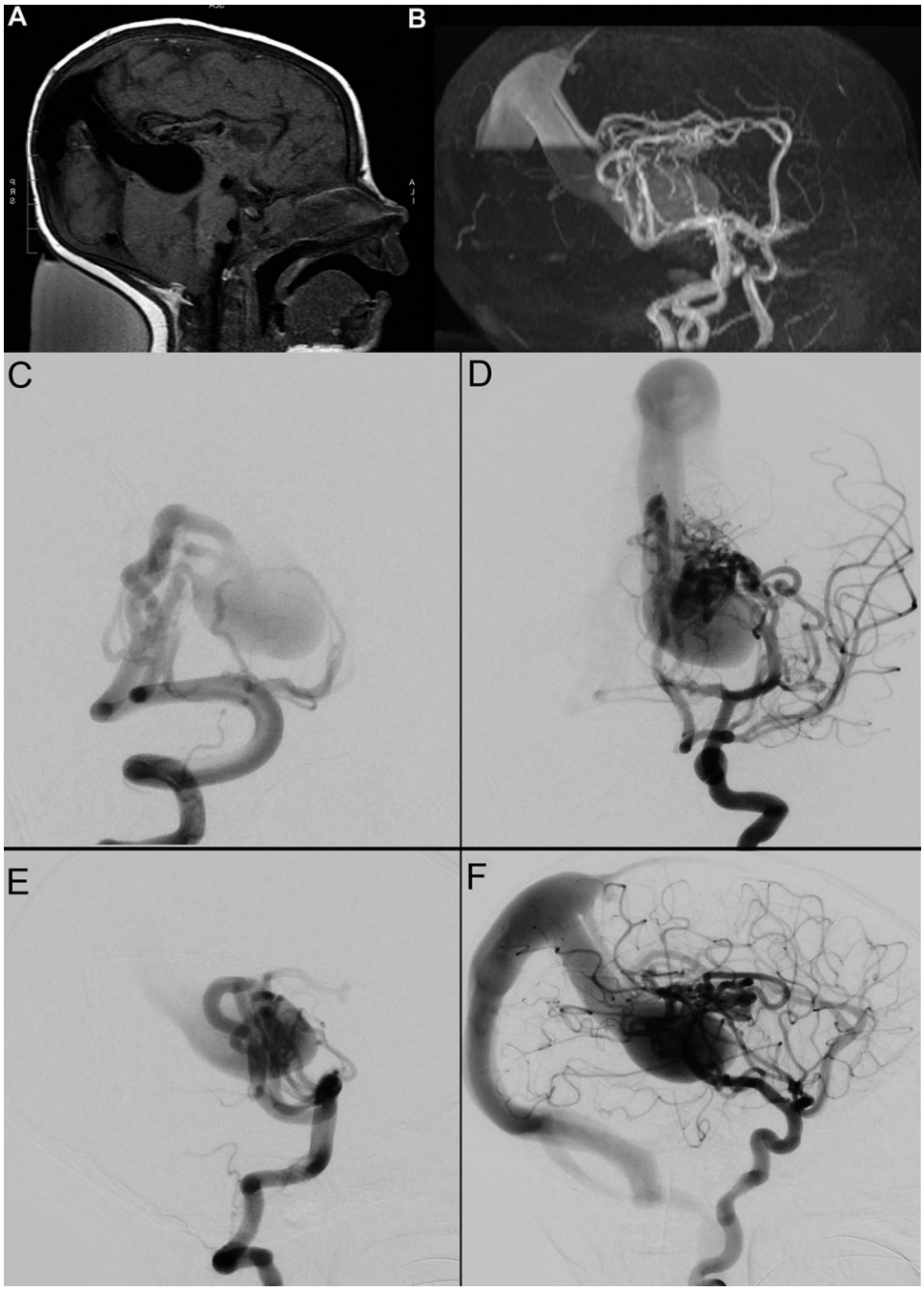

Figure 1 Representative imaging studies. (A) Sagittal T1-weighted MR image demonstrating the dilated prosencephalic vein and persistence of the falcine sinus. (B) Maximum intensity pixel MR angiographic image demonstrating multiple enlarged arterial branches from the anterior and posterior cerebral arteries coalescing on the lateral margins of the dilated recipient vein. (C–F) Composite DSA capturing early (C and E) and delayed (D and F) contrast bolus images. Anterior–posterior (C) and lateral (E) images of a right vertebral arterial injection demonstrate enlarged ipsilateral posterior–lateral choroidal and perimesencephalic branches as well as a smaller thalamoperforating branch coalescing on the anterior–lateral aspect of the recipient vein. Anterior–posterior (D) and lateral (F) images of a left internal carotid artery injection show similarly enlarged posterior–lateral choroidal and perimesencephalic branches arising from the ipsilateral posterior cerebral artery, and smaller branches from the distal pericallosal artery. The lateral projection (F) also demonstrates the dilated prosencephalic vein and persistence of the falcine and occipital sinuses.

In total, 41.2% of 17 cases had radiological evidence of atrophy at baseline and 64.3% of 14 cases had atrophy after treatment. Neither the presence of atrophy at baseline ($p=0.061$) nor after treatment ($p=0.117$) was predictive of functional outcome.

At the last follow-up, the final obliteration of the VOGM was complete in 62.5% of valid cases, partial in 12.5% and incomplete in 25.0% (N=32). Thrombosis on follow-up imaging was achieved in 72.7% of valid cases. White matter calcifications were found in 31.3% of cases on CT. Distal migration of the embolization coils occurred in three cases (7.3%).

DISCUSSION

In this study of 41 children treated for their VOGMs, we have shown that current endovascular embolization procedures allow up to half of patients with the condition to live functionally

Table 3 Treatment approaches and embolic materials employed prior to and after the year 2000. P-values compare between groups treated prior to and following the year 2000.

Total treatments	Total No (%)	Prior to 2000	After 2000	P-value P=0.266
0	1 (2.4)	1 (3.7)	0 (0.0)	
1	10 (24.4)	7 (25.9)	3 (21.4)	
2	16 (39.0)	7 (25.9)	9 (64.3)	
3	5 (12.2)	3 (11.1)	2 (14.3)	
4	3 (7.3)	3 (11.1)	0 (0.0)	
5	4 (9.8)	4 (14.8)	0 (0.0)	
6	1 (2.4)	1 (3.7)	0 (0.0)	
7	1 (2.4)	1 (3.7)	0 (0.0)	
Transarterial approach				P=0.902
0	5 (12.8)	4 (15.4)	1 (7.7)	
1	22 (56.4)	14 (53.8)	8 (61.5)	
2	8 (20.5)	5 (19.2)	3 (23.1)	
3	3 (7.7)	2 (7.7)	1 (7.7)	
4	1 (2.6)	1 (3.8)	0 (0.0)	
Transvenous approach				P=0.604
0	14 (38.9)	9 (37.5)	5 (41.7)	
1	14 (38.9)	8 (33.3)	6 (50.0)	
2	4 (11.1)	3 (12.5)	1 (8.3)	
3	3 (8.3)	3 (12.5)	0 (0.0)	
4	1 (2.8)	1 (4.2)	0 (0.0)	
Transtorcular approach				P=0.076
0	28 (77.8)	16 (66.7)	12 (100.0)	
1	7 (19.4)	7 (29.2)	0 (0.0)	
2	1 (2.8)	1 (4.2)	0 (0.0)	
Embolic agent				
Coils	38 (92.6)	25 (100.0)	13 (100.0)	NA
Silk sutures	8 (19.5)	8 (38.1)	0 (0.0)	P=0.014
NBCA	5 (12.2)	2 (9.5)	3 (25.0)	P=0.233
PVA	5 (12.2)	5 (23.8)	0 (0.0)	P=0.067
Liquid coils	1 (2.4)	0 (0.0)	1 (7.7)	P=0.197
Platinum wire	1 (2.4)	1 (4.8)	0 (0.0)	P=0.425
mRS				P=0.913
0	8 (19.5)	6 (24.0)	2 (18.2)	
1	5 (12.2)	4 (16.0)	1 (9.1)	
2	5 (12.2)	3 (12.0)	2 (18.2)	
3	2 (4.9)	1 (4.0)	1 (9.1)	
4	2 (4.9)	2 (8.0)	0 (0.0)	
5	5 (12.2)	3 (12.0)	2 (18.2)	
6	9 (22.0)	6 (24.0)	3 (27.3)	

p Values for the comparison of groups treated before and after the year 2000.
mRS, modified Rankin Scale; NBCA, N-butyl cyanoacrylate; PVA, polyvinyl alcohol.

normal or near-normal lives. Mixed-type VOGMs were the most commonly seen subtype in our sample. The most commonly observed radiological characteristics included choroidal and posterior cerebral artery feeders, patent falcine veins and sinus stenosis. We showed that poor outcomes were associated with early age of diagnosis, seizures, CHF, and hydrocephalus at presentation, and incomplete embolization. We have also documented changes in the usage of endovascular techniques and embolization materials since 1986.

In our sample, the survival rate after embolization was 78.0%. The overall survival and technical success rates of endovascular treatments for VOGMs from other studies also approach 80%,[11 22–24] suggesting that these approaches are safe and effective. Our observation that many of the children are neurologically normal or near-normal at last follow-up is consistent with previous reports.[10 17 25] Further, early presentation with CHF and seizures was an important predictor of morbidity and mortality in our sample, and the prognosis for patients with VOGMs presenting with CHF in the neonatal period remains poor.[5 6 17] Childhood seizures have been previously associated with cortical injury and poor outcomes, as they may exacerbate neural injury and impair neurodevelopment.[26 27] Therefore, we have confirmed the previously identified markers of prognosis and overall efficacy of endovascular embolization techniques for treating VOGMs.

In contrast to past studies,[28] however, the mixed-type VOGM was the most common subtype in our sample. We did not find choroidal-type VOGMs to be associated with greater functional disability or mortality.[17 25] Perhaps this finding is associated with the use of transvenous embolization approaches at UCSF in cases unsuitable for, or refractory to, transarterial approaches,[29] such as in perinatal, choroidal-type VOGMs. The use of this approach, however, remains controversial.[7 14 30] A major difference between our data and the Bicetre data is that 67/317 (21%) of their patients underwent no angiography/embolization.[10] Since diagnostic angiography was not undertaken by the Bicetre group unless an intervention was planned, the true choroidal:mural ratio for that population is unknown. It is quite possible that patients with choroidal type VOGMs were preferentially not treated at Bicetre because they are sicker at baseline and thus would be excluded from intervention according to that institution's well-described clinical exclusion criteria. The use by our institution of transvenous embolization (our preferred approach rather than choroidal VOGM) may thus reflect our treatment of patients who otherwise would not have been treated at Bicetre. We also favor coil over liquid embolic embolization in patients with heart failure, as there is a lower probability of coil migration to the lungs and thus a smaller chance of worsening heart failure, in contrast to previous studies.[10] Nonetheless, the total number of required procedures is decreasing over time, and the transarterial approach is currently more commonly employed.[25 28] Our rate of complete thrombosis on follow-up imaging in this sample was 62.5%, an improvement on previous reports.[10] Overall, endovascular treatment has been successful at obliterating the VOGM shunts with fewer side effects than neurosurgical approaches.[15 31 32]

The generalizability of our results is limited by the small sample size. We were unable to identify radiological characteristics of some cases directly from imaging because records were not available in 19/41 of our patients. With improvements in electronic medical records and patient tracking, however, we hope that future studies will be able to acquire more complete clinical information on patients with VOGMs, including potentially significant delayed effects of the x-rays used for image-guided therapy.[33] Moreover, the mRS outcome score that we used in this study is not a sensitive measure of patient functional disability and quality of life. Survey-based analyses using standardized instruments may be important in understanding the detailed outcomes of these patients.[17]

We have described a cohort of patients with VOGMs treated with endovascular approaches at UCSF. As these techniques are refined and improved, further understanding of the pathology of VOGMs will guide treatment decisions for each patient. VOGMs continue to be challenging vascular malformations to manage, but neurointerventional approaches allow many children with these malformations to live near-normal lives.

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Contributors MLC performed the chart review, survey analysis, and statistical analysis. She drafted and edited drafts of the article. DLC and SWH planned the study, performed the radiological review and edited drafts of the article. HJF helped in the planning of the study and editing of the article. MRA, JN, CFD, RTH, and VVH performed the endovascular procedures that made the study possible.

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